

Radiology Corner

Angiomyolipoma

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Note: This is the full text version of the radiology corner question published in the December 2007 issue, with the abbreviated answer in the January 2008 issue.

Angiomyolipoma (AML) is a relatively uncommon tumor that can be associated with renal lesions in connection with sporadic or associated tuberous sclerosis disease. These lesions are often found incidentally and managed conservatively with periodic imaging and clinical surveillance, depending on the actual size of the lesion. Lesions greater or equal to 4 cm have been found to have an increased risk for retroperitoneal hemorrhage and may be managed more aggressively. CT scan is currently the imaging modality of choice and the presence of fat within a renal mass is highly suggestive of AML. Other imaging modalities such as ultrasound and MR are also utilized in the evaluation of these renal masses if CT is contraindicated.

History

A 46-year-old male presented to the emergency room with one day history of right flank and right lower quadrant pain. He was mildly hypotensive upon arrival. He was initially evaluated by the emergency room and found to have signs of an acute abdomen. Surgical consultation confirmed rebound tenderness diffusely. An abdominal CT scan was ordered and reviewed, and the patient was taken to the operating room emergently for a right nephrectomy secondary to persistent hemorrhage. Pathology of the tumor confirmed a diagnosis of AML.

Summary of Imaging Findings

Multiple axial abdominal CT scans (figure 1a, 1b, 1c) demonstrate a right heterogeneous lesion coming off the posterior mid-kidney representing a large angiomyolipoma. The surrounding fluid collection represents hemorrhage of the lesion. There was no evidence of left renal involvement.



Fig. 1 (a) Axial abdominal CT scan demonstrates right renal mass and evidence of extra-renal fluid collection indicative of hemorrhage. There is no evidence of left renal lesion or other abdominal organ abnormalities.

Discussion

Angiomyolipoma (AML) is characterized by either a unilateral or bilateral renal mass which can usually be distinguished from other renal masses by the presence of fat found on CT scan. These lesions are almost always benign and many are incidental findings because they are often asymptomatic. AML is associated with isolated sporadic lesions and tuberous sclerosis which is an autosomal dominant disease. Management consists of watchful waiting with intermittent imaging versus surgical intervention for larger or progressive lesions that are symptomatic.

Angiomyolipomas are typically benign neoplasms that may arise from the renal parenchyma and are made up of varying amounts of mature adipose tissue, smooth muscle, and thick-walled vessels (1). This tumor is reported to be found in 0.3 percent of all autopsies and in 0.1 percent of the healthy adult population confirmed via ultrasound and CT scan (2). It was initially discovered by Fisher in 1911 and was later designated Angiomyolipoma by Morgan in 1951. AML is derived from perivascular epithelioid cells and its growth is proposed to be hormone related because of its female predominance and rare incidence before puberty. Pregnancy is also considered a risk factor associated with these lesions (3).

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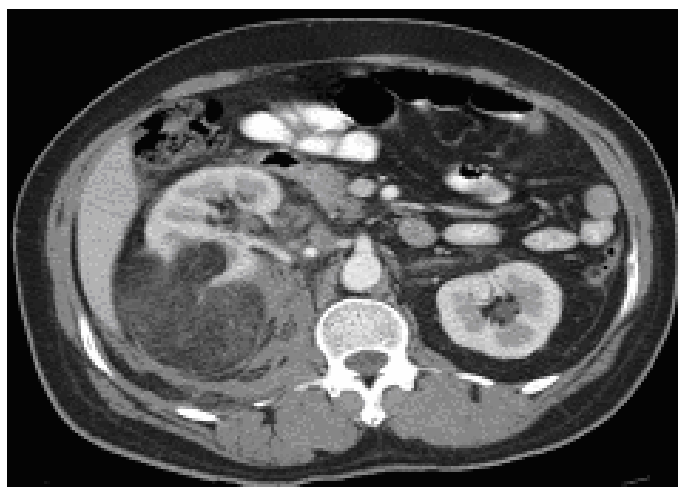


Fig. 1 (b) Axial abdominal CT scan demonstrates ruptured lesion and hemorrhage.

AML is seen in two clinical settings to include sporadic or isolated, and tuberous sclerosis. Sporadic AML accounts for approximately 80% of cases and is more common in women with a 4:1 ratio and a mean age of 43 years. The classic clinical presentation of AML may be flank pain on the affected side of the lesion, a palpable and tender renal mass, and gross or microscopic hematuria (4). Sporadic AML is often an incidental finding when imaging is obtained for other clinical indications. Patients with sporadic disease tend to present later in life with smaller tumors and a lower incidence of bilateral renal involvement in comparison to AML associated with tuberous sclerosis (4).



Fig. 1 (c) Axial CT scan shows extent of hemorrhage at lower kidney pole.

Tuberous sclerosis (TS) is an autosomal-dominant disorder characterized by mental retardation, epilepsy, and a distinctive skin lesion known as adenoma sebaceum (aka “tubers”). This condition is also associated with a high incidence of angiomyolipoma. TS accounts for approximately 10-20% of AML cases with a female-to-male predominance of 2:1 and a mean age of 30 years (4). TS can also be associated with other renal conditions to include benign cysts and lymphangiomas. The prevalence of AML in these patients

increases with age as demonstrated by a study of 60 children who underwent serial ultrasounds in which 55 percent of children with TS had a renal lesion at the mean age of 6.9 years, which increased to a reported 80 percent at a mean age of 10.5 years (5). Similar to sporadic cases of AML, AML lesions associated with TS are often asymptomatic. TS can be associated with more aggressive AML lesions that become large and lead to complications such as abdominal and flank pain, and even bleeding. In some cases, progressive AML in these patients leads to end-stage renal disease secondary to replacement and compression of renal parenchyma via mass effect (6).

CT scan is currently the most useful radiologic tool to diagnose AML, and the presence of fat within a renal lesion confirmed by an attenuation value of -15 Hounsfield units is suggestive of AML (7). Renal MRI may be necessary to help determine the character of more complex or smaller lesions in addition to CT. It is important to evaluate the relationship of the fat to the remainder of the tumor to assess whether it is intratumoral and not perirenal, which could be indicative of an expanding renal cell carcinoma. In rare instances, an RCC may show fat attenuation on CT scan and is caused by entrapment of the perineal sinus fat, lipid necrosis, or osseous metaplasia (8). Although AML is usually not associated with a potential for malignancy, there was a recent case report that demonstrated the development of adult renal cell carcinoma in a 40 year-old woman with preexisting AML, which demonstrates a rare potential that RCC might arise within AML (9). Other case reports discuss the potential for angiomyolipoma to mimic malignant lesions from a radiological perspective, which can present a challenge to pursue medical versus surgical intervention (10). Ultrasound and MR imaging may also be useful in the evaluation of these renal masses, especially if there is a contraindication to CT. Ultrasound usually demonstrates a well circumscribed, reflective mass that is more echogenic than central sinus fat. This increased echogenicity is secondary to the fat content, multiple interfaces, heterogeneous cellular architecture, and multiple vessels within the tumor. Ultrasound also shows posterior shadowing while a small renal cell carcinoma often shows a hypoechoic rim and intratumoral cystic changes (11).

As mentioned above, AML is often found incidentally and lesions less than 4 cm are managed conservatively with periodic imaging and clinical follow up. Lesions greater than 4 cm are associated with an increased risk of hemorrhage and may be treated surgically with elective nephrectomy. Wunderlich's syndrome is massive retroperitoneal hemorrhage from AML and is a feared complication in up to 10% of patients with this condition (12). Selective embolization and partial nephrectomy are other treatment modalities that may be performed in an attempt to preserve kidney function.

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